

## Hypoplastic Left Heart Syndrome (HLHS)

The aim of this fact sheet is to explain what Hypoplastic Left Heart Syndrome is, what effect it will have on a child and how it can be treated.

### What is Hypoplastic left heart syndrome?

Hypoplastic means 'not developed'. Left heart means the structures on the left side of the heart. A syndrome is a number defects together. So hypoplastic left heart syndrome (HLHS) means that the left side of the heart has not developed properly.

The left ventricle, which should pump red (oxygenated) blood to the body, is very small, and there is a complete blockage between it and the aorta. The aorta, which should carry red blood to the body, is much too small.

Fig 1 – Hypoplastic Left Heart Syndrome

#### Hypoplastic left heart syndrome

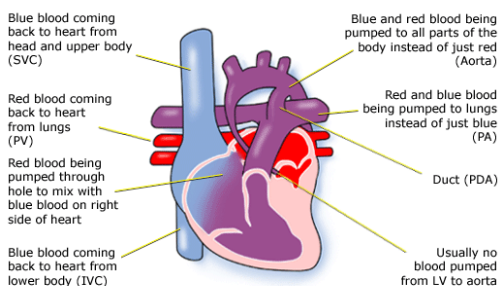
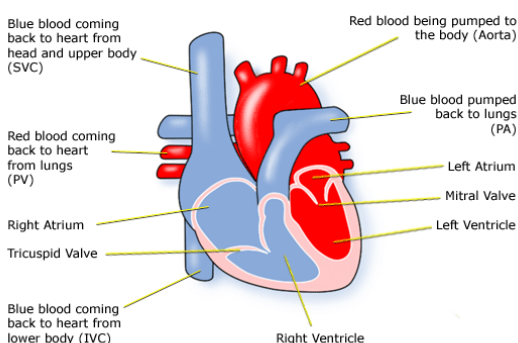


Fig 2 – Normal Heart

#### Normal Heart



### Diagnosis

Your baby may have been diagnosed as having a heart condition before birth. HLHS may be seen on a scan of the foetal heart. HLHS can develop if there is a blocked or narrow valve, when the left side of the heart does not grow as it should during the later stages of pregnancy.

After birth, your baby can survive while the foetal circulation system (ductus arteriosus) is still open. This is because red blood will still be crossing from the left atrium to the right. From there it will be pumped, with blue blood, into the pulmonary artery. While the ductus arteriosus is still open, blood will flow through it into the aorta, and thus round the body.

Soon after birth, however, the foetal circulation will start to close down. Your baby will become extremely ill as less and less oxygen reaches the organs. They may look pale and blue (cyanosis) and have difficulty breathing and feeding. Most babies will need treatment within the first days or months of life.

When a heart condition is detected the tests used can be:

- pulse, blood pressure, temperature, and number of breaths a baby takes a minute
- listening with a stethoscope for changes in the heart sounds
- an oxygen saturation monitor to see how much oxygen is getting into the blood
- a chest x-ray to see the size and position of the heart
- an ECG (electrocardiogram) to check the electrical activity an ultrasound scan (echocardiogram) to see how the blood moves through the heart

- checks for chemical balance in blood and urine
- a catheter or Magnetic Resonance Imaging test may be needed.

## Treatment

HLHS is a very difficult problem to treat. The treatment for this condition is palliative. This means your child's heart cannot be corrected, that is, made to work like a normal heart. But in some cases it can be improved with a procedure called the Norwood operation.

### Norwood Procedure

This is a high risk procedure. It is a series of three surgical operations which would eventually allow the right side of your child's heart to take over the work of the left side. The aim of the three operations is for the right ventricle to pump red blood to the body, while the blue blood is allowed to flow directly to the lungs.

### Stage One

In the first few days of life, the wall between the left and right atrium is removed so that red blood coming back from the lungs will pass into the right atrium, and from there to the right ventricle. The pulmonary artery is attached to the aorta. A shunt (passage) is created between the aorta and the pulmonary artery branches to the lungs. Mixed red and blue blood will now be pumped through the pulmonary artery to the aorta, to both lungs and to the body.

### Stage Two

Between the ages of four to nine months the blood flow to the lungs is increased. The SVC (superior vena cava), which carries blue blood from the top of the body to the right atrium, is joined directly to the pulmonary arteries. The shunt between the aorta and the branch arteries created at stage one is closed.

### Stage Three

The IVC (Inferior vena cava), which carries blue blood from the lower part of the body to

the right atrium, is joined directly to the pulmonary arteries. This procedure is also known as the Fontan operation.

There is a risk to your child in all the procedures, but how great that risk is depends on the shape of the individual heart, and how well your child is otherwise. The doctors will discuss risks with you in detail before asking you to consent to any of the operations.

## Other forms of treatment

### Transplantation

Some doctors will recommend a heart transplant to treat HLHS. This will involve trying to keep your baby's foetal circulation open, using medicines or a stent in the ductus arteriosus, while waiting for a heart to become available. At present, however, transplantation is rarely available in the UK as there are very few baby size hearts available.

## How your child is affected

Treatment for HLHS is very complicated - there cannot be any guarantees of how well your child will do during or after surgeries.

You may notice some of the following things after your child's treatment.

- ♥ Your child may have fluid in the lungs (pleural effusion) after surgery. It is also not uncommon for a child to pick up an infection, such as a chest infection or infected wound, while undergoing treatment. There may be other complications which your cardiologist should explain to you.
- ♥ Scars on the chest fade very rapidly in most children, but they will not go altogether. Smaller scars on the hands and neck usually fade away to nothing.
- ♥ Some children react badly to some kinds of medicines.

- ♥ Many children with this condition will continue to have a bluish tinge to their skin (cyanosis).
- ♥ Heart valves and shunts may need further surgery as the child gets older and bigger.
- ♥ Some children develop a very fast heart rate (called tachycardia) or a slow heart rate (bradycardia). They may need medication to keep it stable, or a pacemaker to correct it.
- ♥ Hearts that are not normal are more likely to have an infection called infective endocarditis. Although rare this is a difficult disease to treat. Read more about infective endocarditis and how to prevent it in our fact sheet 'Infective endocarditis' – order by calling our freephone infoline 0808 808 5000 or download from our website [www.chfed.org.uk](http://www.chfed.org.uk).
- ♥ Because the flow of blood is slower than it should be, there may be a danger of blood clotting. If the doctors think this is a

concern, your child may need to take aspirin or warfarin. Aspirin and warfarin are anticoagulant medicines (stop blood cells sticking together). If your child is on warfarin, they will need to have their blood tested regularly. Read more in our fact sheet 'Warfarin' - order by calling our freephone infoline 0808 808 5000 or download from our website [www.chfed.org.uk](http://www.chfed.org.uk).

Your child may have other health problems as well. Seek advice about claiming Disability Living Allowance (DLA) if your child needs much more care than another child of their age, or if they are having problems walking.

The long term outcomes for children are not yet known.

### More information

Little Hearts Matter is the UK support group for families of children who have this disorder – contact them on 0121 455 8982 or visit their website [www.lhm.org.uk](http://www.lhm.org.uk).